CASE REPORT

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Agenesis of the infra-hepatic segment of the inferior vena cava associated with recurrent deep venous thrombosis: case report

Agenesia de segmento infra-hepático de veia cava inferior associada a trombose venosa profunda de repetição: relato de caso

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Abstract

Agenesis of the inferior vena cava (IVC) has been described in less than 1% of the population; a rare occurrence caused by embryonic abnormalities. Its correlation with deep vein thrombosis (DVT) is certainly underestimated, since this change is hard to detect using ultrasound. The aim of the article is to report the case of a 41-year-old female patient with pain and edema up to the top of the right thigh after plastic surgery. Bilateral venous duplex ultrasound revealed bilateral DVT involving iliac-femoral-popliteal and distal segments. Venous angiotomography was requested because the IVC was not visible on ultrasound, revealing thrombosis of the right lumbar plexus and iliofemoral segment bilaterally and agenesis of the infrahepatic segment of the inferior vena cava, with ectasia and compensatory tortuosity of paravertebral veins and the azygos-hemiazygos system, and bilateral pelvic varices. Systemic and oral anticoagulation were administered, with a satisfactory clinical response.

Keywords: inferior vena cava; deep venous thrombosis; cardiovascular abnormalities.

Resumo

A agenesia de veia cava inferior é descrita em menos de 1% da população, de ocorrência rara e devido a alterações embrionárias. Sua correlação com a trombose venosa profunda certamente é subestimada, visto que tal alteração é de difícil detecção pela ultrassonografia. O objetivo deste artigo foi relatar o caso de uma paciente de 41 anos com dor e edema até a raiz de coxa direita após cirurgia plástica. Foi realizado dúplex venoso de membros inferiores e evidenciada trombose venosa profunda ilíaco-femoro-poplíteo e distal bilateralmente. Solicitado angiotomografia venosa devido a não visualização de veia cava inferior no ultrassom, evidenciando trombose de plexo lombar direito e segmento ilíaco-femoral bilateral, além de agenesia de segmento infra-hepático de veia cava inferior, com ectasia e tortuosidade compensatória de veias paravertebrais, sistema ázigos e hemiázigos, com varizes pélvicas bilateralmente. Foi realizada anticoagulação sistêmica e oral, com boa evolução clínica.

Palavras-chave: veia cava inferior; trombose de veia profunda; anomalias cardiovasculares.

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INTRODUCTION

The most common cause of agenesis of the inferior vena cava (IVC) during embryogenesis is dysgenesis with a venous system defect, which may be secondary to intrauterine or perinatal thrombosis, without accompanying embryonic abnormalities.[1] Anomalies of the IVC have been diagnosed incidentally with greater frequency since the advent of imaging exams, since the majority of patients are asymptomatic.[2-5]

Deep venous thrombosis (DVT) is caused by a congenital or acquired hypercoagulable state. Absence of the IVC may be a risk factor, since it impairs drainage of blood from the lower limbs and causes a state of venous stasis and hypercoagulability, in which venous return is dependent on a system made up of collaterals.[1-3,6-10] This study was duly evaluated and approved by a Research Ethics Committee (CAAE 45875521.4.00000.5411 and ruling number 4.699.406).

DESCRIPTION OF THE CASE

A 41-year-old female patient presented at a vascular surgery service with pain of moderate intensity in the right lower limb, associated with accentuated edema around the hip. Six days earlier she had undergone abdominal liposuction, mastopexy, and breast augmentation with implants.

She had no comorbidities and had been taking combined oral contraception with levonorgestrel 0.15 mg and ethinylestradiol 0.03 mg for the preceding 1 year and 6 months, which was withdrawn after diagnosis since it is a risk factor. She had a personal history of DVT in the left lower limb, in the femoral-popliteal segment, at 22 years of age, during postpartum after a natural delivery, when she had been treated with anticoagulation with a vitamin K antagonist (warfarin) for 1 year, having been discharged to outpatients follow-up after treatment.

Clinical assessment found her in good general health, with no changes to vital signs, with discrete abdominal edema and minor ecchymosis, primarily in the pubic area, with small quantities of serous secretions from the surgical wound in the right iliac fossa. Lower limbs were free from cyanosis and pallor, with moderate bilateral edema, more pronounced on the right, and with palpable distal pulses.

The results of laboratory tests requested at admission included: hemoglobin 11.1 g/dL (normal range [NR]: 13.5-17.5 g/dL), hematocrit 32.3% (NR: 41-53%), platelets 136,000 µL (NR: 140,000-440,000 µL), and white blood count 11,700/µL (NR: 4,000-11,000/µL). Homocysteine, urea, creatinine, albumin, transaminases, coagulogram, and electrolytes were all normal. As such, the patient's laboratory tests revealed mild anemia, thrombocytopenia, and leukocytosis.

Vascular ultrasonography with duplex (USD) was used to examine the deep vein system of the lower limbs and the abdominal venous system. These examinations revealed acute DVT in the iliac-femoral-popliteal segment and distal segments bilaterally. The IVC was not visible in the USD examination, and, in view of the patient's postoperative status with edema of the abdominal subcutaneous tissues, an additional imaging exam was conducted for diagnostic confirmation. Venous angiotomography of the abdomen and pelvis identified venous thrombosis of the right lumbar plexus (Figure 1) and of the iliofemoral segment bilaterally (Figure 2) and agenesis of the infra-hepatic segment of the IVC (Figure 3), which originated at the confluence of the renal veins (Figure 4), with associated compensatory ectasia and tortuosity of the left gonadal vein, paravertebral veins, and the azygos-hemiazygos system (Figure 5), with pelvic varicose veins bilaterally.

The patient was initially treated with systemic anticoagulation in hospital, using unfractionated heparin via a continuous infusion pump at an initial dose of 18 UI/kg/h. The pain and edema in her lower limbs gradually diminished and she was discharged from hospital on the seventh day, with a prescription

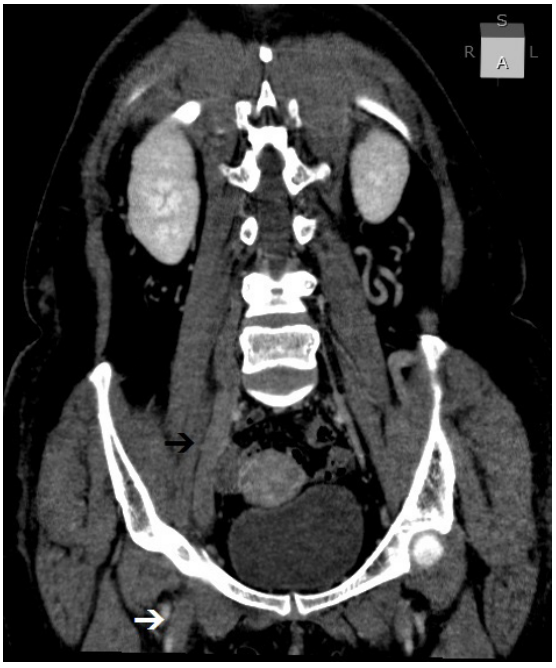

Figure 1. Venous angiotomography, coronal anterior oblique view, showing venous thrombosis of the right common femoral vein (white arrow) and the right lumbar plexus (black arrow).

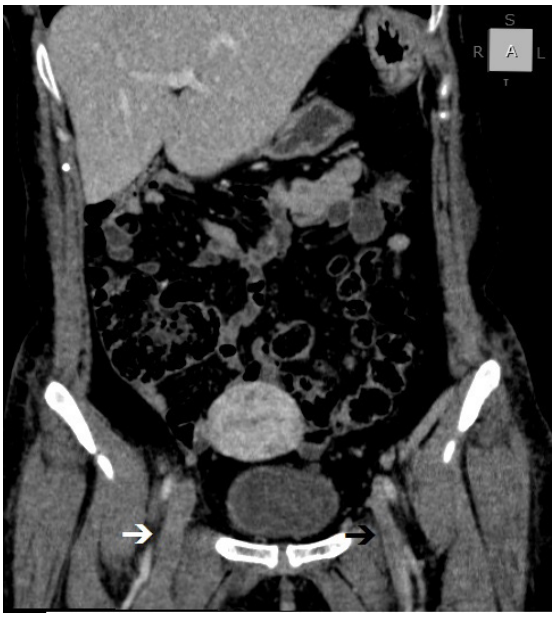

Figure 2. Venous angiotomography, coronal view, showing right (white arrow) and left (black arrow) iliofemoral venous thrombosis.

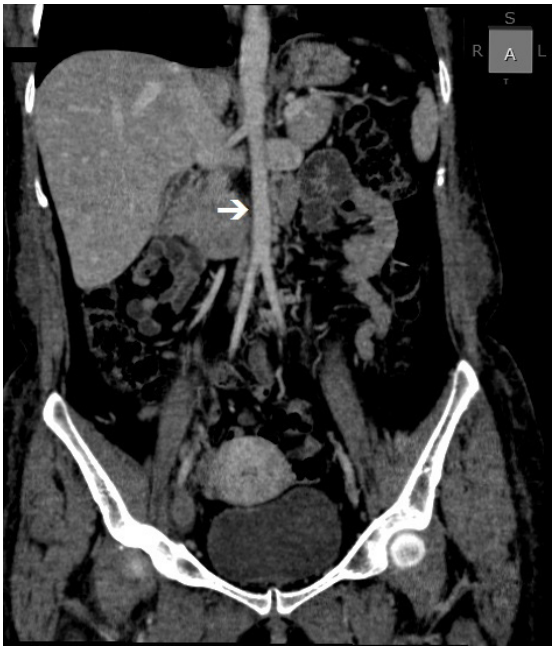

Figure 3. Venous angiotomography, coronal view, revealing agenesis of the infra-hepatic segment of the inferior vena cava. Note the abdominal aorta with no adjacent vascular structure (arrow).

for indefinite oral anticoagulation with warfarin and instructions to adopt the Trendelenburg position when lying supine, wear mid-thigh medium compression elastic stockings (20-30 mmHg), and attend outpatients

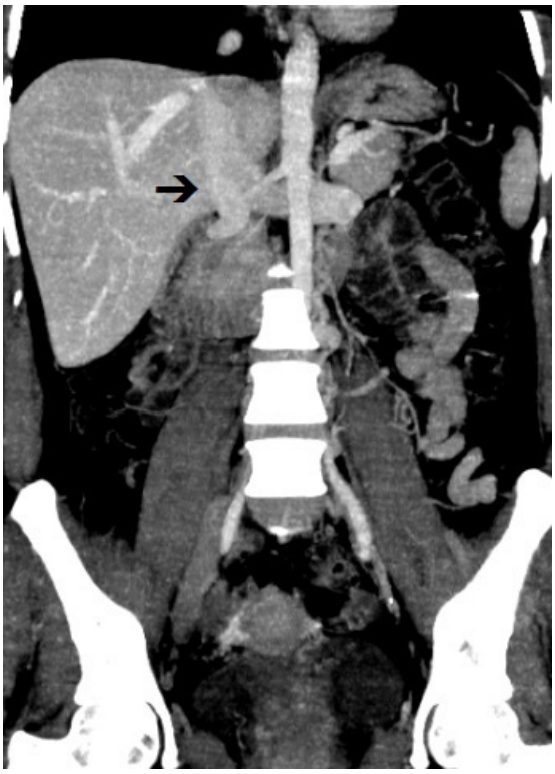

Figure 4. Venous angiotomography, coronal view, showing the origin of the inferior vena cava from the confluence of the renal veins (arrow).

follow-up. At periodic outpatients consultations (14, 30, and 60 days and every 90 days thereafter), she was free from complaints, her anticoagulation level was within the proposed target range (prothrombin activity time 2.5-3.5), symptomology had improved, and she had had no bleeding episodes.

DISCUSSION

Embryological formation of the IVC is a complex process involving many anastomoses of pairs of embryonic veins.[2-4,11] Development of the infra-hepatic segment of the IVC starts between the sixth and eighth weeks of intrauterine life, and several theories as to its formation have been proposed. Normally, the IVC comprises four segments: the hepatic, derived from the vitelline vein; the suprarenal, which develops from the right subcardinal vein; the renal, from the right supracardinal vein; and the infrarenal, which also derives from the right supracardinal vein.[4-6]

Congenital anomalies of the IVC have estimated prevalence de 0.07% to 8.7% in the general population.[2] Many different anomalies have been described, including an IVC on the left, a double IVC, continuation of the

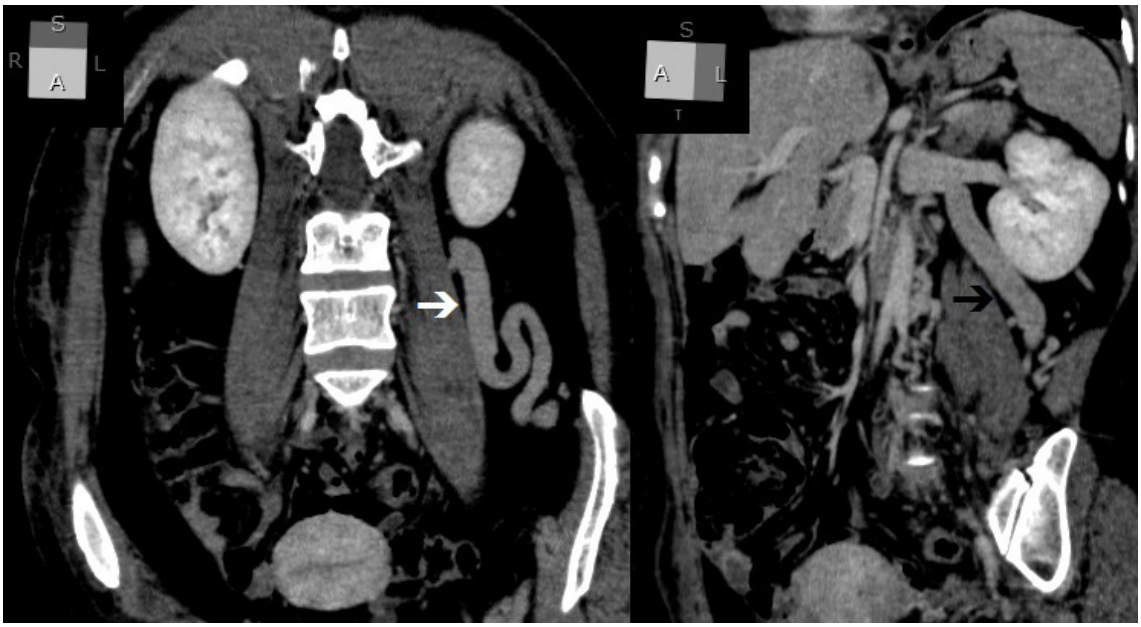

Figure 5. Venous angiotomography, coronal view (left) and coronal oblique view (right), showing tortuosity (white arrow) and ectasia of the left gonadal vein (black arrow).

IVC to the azygos, a circum-aortic left renal vein, a retroaortic left renal vein, a circum-caval ureter, agenesis of the hepatic segment of the IVC, and infrarenal agenesis with preservation of the suprarenal segment, as described in the present report.[4,12] Agenesis is described in less than 1% of the population; a rare occurrence that is caused by abnormal embryonic development.[7,13] The majority of cases of partial absence of the IVC affect its supra-hepatic portion (90%) and there are associations with congenital heart disease in 0.6% to 2% of cases or with other cardiac anomalies in 0.3% to 0.5%.[14] Agenesis of the infrarenal segment of the IVC is extremely rare, considering that only 6% of these anomalies involve the renal or infrarenal segments.[13]

It has been suggested that agenesis of the IVC should be considered in young patients with proximal DVT that is idiopathic, bilateral, and recurrent, in the absence of predisposing risk factors such as thrombophilia and especially in patients under the age of 30.[1-3,7,8,10,11,13,15,16] Known risk factors for DVT also have an influence, acting in synergy if combined with IVC agenesis. These include genetic factors that lead to hypercoagulability, such as deficiencies of proteins C and S and antithrombin; factor V Leiden, high concentrations of factor VIII and hyperhomocysteinemia, in addition to acquired risk factors such as traumas, surgery, immobilization, and pregnancy.[9,10] Other thrombogenic factors described include intense muscle exercise, long journeys, and oral contraceptives.[2,3]

The association between IVC agenesis and DVT is undoubtedly underestimated, since this anomaly is unlikely to be detected with USD and other diagnostic methods such as computed tomography or angiography are needed when there is a suspicion of involvement of supra-inguinal segments.[8,15,16]

Knowledge of anatomic variations is important to avoid diagnostic errors and so that surgeons are aware of possible intraoperative complications, such as ligation of collateral veins with serious pathophysiologic consequences or even death.[4,6,13,14] Patients who are diagnosed with these vascular anomalies should be advised to avoid thrombogenic risk factors because of the high risk of thrombosis and recurrence.[2,10]

To date, there are no reports giving indications for interventional treatments for IVC agenesis and there is insufficient data to recommend prophylactic treatment of these patients. There are also no indications with regard to prolonged use of anticoagulants in patients who have had DVT previously.[10,11,13]

CONCLUSIONS

Diagnostic suspicion is indispensable, particularly in young patients with recurrent proximal DVT, in order to proceed with correct etiologic diagnosis, anticoagulant therapy, and patient guidance. With

regard to duration of treatment and the therapeutic target, controlled studies should be conducted to guide conduct in the future.

REFERENCES

1. Gensas CS, Pires LM, Kruse ML, Leiria TLL, Gomes DG, de Lima GG. Agenesia da veia cava inferior. Rev Bras Cardiol Invasiva. 2012;20(4):427-30. http://dx.doi.org/10.1590/s2179-83972012000400015.
2. Obernosterer A, Aschauer M, Schnedl W, Lipp RW. Anomalies of the inferior vena cava in patients with iliac venous thrombosis. Ann Intern Med. 2002;136(1):37-41. http://dx.doi.org/10.7326/0003-4819-136-1-200201010-00009. PMid:11777362.
3. Gayer G, Luboshitz J, Hertz M, et al. Congenital anomalies of the inferior vena cava revealed on ct in patients with deep vein thrombosis. AJR Am J Roentgenol. 2003;180(3):729-32. http://dx.doi.org/10.2214/ajr.180.3.1800729. PMid:12591684.
4. Bass JE, Redwine MD, Kramer LA, Huynh PT, Harris JH Jr. Spectrum of congenital anomalies of the inferior vena cava: Cross-sectional imaging findings. Radiographics. 2000;20(3):639-52. http://dx.doi.org/10.1148/radiographics.20.3.g00ma09639. PMid:10835118.
5. Kellman GM, Alpern MB, Sandler MA, Craig BM. Computed tomography of vena caval anomalies with embryologic correlation. Radiographics. 1988;8(3):533-56. http://dx.doi.org/10.1148/radiographics.8.3.3380993. PMid:3380993.
6. Viana SL, de Mendonça JLF, Freitas FMO, et al. Anomalias congênitas da veia cava inferior: valor dos métodos multiplanares em seu diagnóstico: ensaio iconográfico. Rev Imagem. 2006;28(4):233-9.
7. Lichtenfels E, Becker AS, Pires VC, Bonamigo TP. Diagnóstico etiológico da trombose venosa profunda de repetição dos membros inferiores. J Vasc Bras. 2006;5(2):157-9. http://dx.doi.org/10.1590/S1677-54492006000200014.
8. Ruggeri M, Tosetto A, Castaman G, Rodeghiero F. Congenital absence of the inferior vena cava: a rare risk factor for idiopathic deep-vein thrombosis. Lancet. 2001;357(9254):441. http://dx.doi.org/10.1016/S0140-6736(00)04010-1. PMid:11273066.
9. Basile A, Certo A, Ascenti G, Lamberto S, Cannella A, Garcia Medina J. Embryologic and acquired anomalies of the inferior vena cava with recurrent deep vein thrombosis. Abdom Imaging. 2003;28(3):400-3. http://dx.doi.org/10.1007/s00261-002-0089-0. PMid:12719913.
10. Konopka CL, Salame M, Padulla GA, Muradás RR, Batistella JC. Agenesis of inferior vena cava associated with deep venous thrombosis. J Vasc Bras. 2010;9(3):196-9. http://dx.doi.org/10.1590/S1677-54492010000300019.
11. Alves EC, Ferro GBR, França LKL, Jacó MB, Pitta GBB. Ausência de veia cava inferior: relato de caso. J Vasc Bras. 2010;9(4):254-6. http://dx.doi.org/10.1590/S1677-54492010000400010.
12. Yang C, Trad HS, Mendonça SM, Trad CS. Anomalias congênitas da veia cava inferior: revisão dos achados na tomografia computadorizada multidetectores e ressonância magnética. Radiol Bras. 2013;46(4):227-33. http://dx.doi.org/10.1590/S0100-39842013000400009.
13. Onzi RR, Costa LF, Angnes RF, et al. Malformação de veia cava inferior e trombose venosa profunda: fator de risco de trombose venosa em jovens. J Vasc Bras. 2007;6(2):186-9. http://dx.doi.org/10.1590/S1677-54492007000200016.
14. Felicio ML, Martins AS, Andrade RR, Silva MADM. Ausência parcial de veia cava inferior associada à malformação intestinal. Rev Bras Cir Cardiovasc. 2007;22(3):362-4. http://dx.doi.org/10.1590/S0102-76382007000300017. PMid:18157426.
15. Langer F, Santos D, Suertegaray G, Haygert C. Bilateral deep vein thrombosis associated with inferior vena cava agenesis in a young patient manifesting as low back pain. Acta Med Port. 2017;30(4):333-7. http://dx.doi.org/10.20344/amp.7744. PMid:28555560.
16. Faria JP No, Sousa Miranda RL. Agenesia de veia cava inferior associada à trombose venosa profunda em jovens. Rev Uningá. 2018;55:35-41.

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